

THE OPERATIVE TREATMENT OF NON-MICRO- CEPHALIC IDIOCY.

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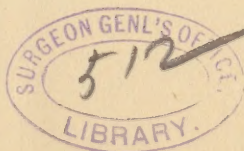
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WITHIN the past few years much attention has been paid to the surgical treatment of various cerebral diseases, which had formerly been considered incurable, and the sufferers from which either had to be relegated to some institute for the care of imbeciles or remain a burden in their homes, requiring constant watching and care. To Fuller, of Grand Rapids, Mich., belongs the credit of having first suggested the idea that in cases of imbecility accompanied by microcephalus something might be effected by relieving cerebral tension. Much has now been written on this subject, but little attention seems to have been paid to cases of imbecility in which microcephalus was absent or not marked.

As I have operated on two cases belonging to the latter category,—one fatal and the other not,—and have had the pleasure of examining and observing some other cases, it may possibly be of some interest to review these two cases, and make a few remarks on the subject of imbecility in general, without microcephalus.

CASE I.—*Imbecility consequent to an attack of Meningitis; Craniotomy; Death.*—L. H., female, age four years, Kansas City, Kan., was brought to me in the end of March, 1892. When patient was eight months old she could say a few words,—e.g., “papa,” “mamma,” etc. About that time she suffered from meningitis, being unconscious for a period of four or five days. From this illness patient’s imbecility dated. Nine weeks ago a similar attack of meningitis was passed through. Patient can now say nothing but “papa.” She will



not play with other children, is mischievous, and given to knocking her head against various objects. The calls of nature are not attended to. During the past ten days she has been crying a great deal. The head appears well developed, especially posteriorly, the development along the coronal sutures being less than elsewhere. In spite of a gloomy prognosis as to the immediate danger of an operation, and the statement that the work would be purely experimental, the parents desired me to operate. My judgment as to the justifiability of an experimental operation in this case was based (1) on the results often obtained by what White terms "operations *per se*" in epilepsy; (2) on the results obtained by craniotomy in cases of microcephalus.

On April 2, 1892, assisted by Drs. Lanphear and Thrush, I operated in All Saints' Hospital. The operation consisted in removing, with trephine and Keen's forceps, a wide strip of bone from a point slightly behind the vertex to the edge of the hairy scalp in front. The left side was first attacked. The skull was found to be $\frac{1}{8}$ inch thick posteriorly and $\frac{3}{16}$ anteriorly. When first exposed the dura did *not* pulsate, but pulsation soon appeared. The dura was thick, hard, and in the posterior part of the exposed area somewhat injected. It was very firmly attached to the bone. On the right side dural pulsation was found as soon as the skull was opened. Immediately after operation shock was great, but fair reaction was obtained before long. The temperature on the evening of the operation was 101° F., but by four o'clock next morning it had run up to 106°, while the pulse was fair, though rapid. The skin was dry and the face cold. At 8 A.M. the patient died. No autopsy could be obtained. I regret exceedingly having operated on both sides of this head at one sitting, as, had the operation been confined to one side at one sitting, probably the child would have survived the injury.

CASE II.—*Imbecility probably due to Maldevelopment of Cortex Cerebri; Craniotomy; Improvement.*—A. B., male, aged twelve years, son of healthy parents, with good family history. The patient was brought to me on December 2, 1892. When four months old he fell from his nurse's knee, but whether he sustained any injury or not is unknown. Up to this time he had shown no signs of any mental want. Between the ages of nine and twenty months he suffered from "fits." During this period, and for years afterwards, he would not notice anything. Walking began when he was two or three years of age. Patient was deaf up to his sixth year, but has now very acute hearing.

Present Condition.—The patient is a thin but apparently healthy boy of cleanly habits; he is extremely restless; when standing he continuously sways from side to side, standing first on one foot and then on the other; his hands are in constant but apparently voluntary motion; his face uniformly presents a worried, puzzled expression, and he constantly frowns; the attention is never fixed on anything for more than a few seconds; he seems to know what is said to him, but won't answer questions; if he knows a certain article and where it lies, he will fetch it when asked to do so, but does not understand explanations as to what is wanted or where it is; he is generally obedient, though often stubborn; he is very musical, and can hum any tune he happens to hear; sad music affects him much.

The head is not microcephalic, but there is a slight depression near the coronal suture on both sides, more marked on the left than the right.

January 7, 1893. I operated with the assistance of Drs. Lilly and Thrush. The skull on the left side was exposed by a long antero-posterior flap, the base of which was anterior and corresponded to the edge of the hairy scalp; a trephine was applied to the posterior portion of the frontal region one inch from the middle line; the button of bone removed was of normal thickness; the dura bulged into the opening, but *no* pulsation was present; with Keen's forceps a strip of bone three and a quarter inches long and varying in width from one-half to one inch was removed; the bone was thickest (one-fourth inch) where there was a depression beside the coronal suture; hæmorrhage from the diploë was free; pulsation was now visible and palpable; the dura was opened at the point where the trephine had been applied, and about one drachm of clear fluid escaped; the surface of the brain appearing healthy, the wound in the dura was sutured and the scalp replaced; the morning following the operation the patient's relatives were astonished to hear him ask for a drink of water, a thing previously unknown in his history. From that day on he asked for various things he desired, such as "cracker," "orange," etc., and when, in spite of his struggles, it was sought to give him a dose of calomel, he very vigorously protested by saying, "Stop that."

On January 27 he named me correctly for the first time.

On April 17 I operated on the *right* side, assisted by Drs. Griffith and Lilly. The method employed was the same as already described. The parietal bone was found to be very red in color, while the frontal

was pale ; when the skull was opened pulsation was very feeble, but it soon became more vigorous ; the brain bulged into the opening, especially opposite the depression on the skull already referred to ; on the evening after the operation the patient was extremely thirsty, but the attendants, in accordance with my instructions, abstained from giving him much fluid ; when I saw him he was constantly calling for every sort of thirst-satisfier he knew, and so I ordered the nurse to give him there and then some ice ; soon after I left he again became thirsty, and demanded "ice," "water," "pop-water," "orange," etc. ; the nurse being obdurate, he called for me repeatedly by name. I mention this to show that the patient had become capable of expressing his wants and of *reasoning* that, as I had given him ice before, I might allow him to have it again, if I was present. When I last saw the patient, a few days ago, he recognized me by name, although he had not seen me for several months. His family, who are exceptionally intelligent people, consider him infinitely happier than formerly ; he has lost his frowning, worried expression ; none of his early gains have been lost, but under his sister's tuition distinct progress has been obtained ; he will frequently answer simple questions correctly, and asks for many things which he desires ; in various ways he gives evidence of a capability for reasoning. Although he is still an imbecile, yet he is not such a burden to his relatives as he formerly was, and there are signs that a capability for education exists which was formerly entirely absent.

In the first case which is here reported, to what cause must death be ascribed ? It must have been due to one of three causes,—viz.: (1) Shock ; (2) meningitis ; (3) hyperpyrexia due to injury of certain thermotaxic centres. The history of the reaction and the great elevation of temperature disposes of the shock theory. One would hardly expect meningitis to develop and cause death within a period of less than twenty hours after an operation, even although the patient presented the history of having previously suffered from meningitis. To my mind the third explanation appears the most plausible. It is based on observations made by Mr. Horsley and on cases reported to me by two of my colleagues in Kansas City. In one fatal case of craniotomy, reported by Horsley,¹ after a careful autopsy had been made, he was forced

¹ Brit. Med. Journ., 1891, Vol. II, p. 580.

to the belief that hyperpyrexia caused by injury to certain cortical centres was the cause of death, and that these centres were in the motor region. His conclusion as to the existence of such centres was fortified by the observation that in various forms of paralysis due to cortical lesions the temperature of the paralyzed limbs was higher than that of their fellows on the opposite side. Dr. Lanphear informs me that in a case in which he trephined and subsequently used drainage with a rubber tube, the temperature suddenly ran up to $104\frac{1}{2}^{\circ}$ F. On opening the wound, it was found that the drainage-tube had slipped under the skull and lay between dura and bone over the motor area. There were no signs of meningitis. Removal of the offending drain was quickly followed by a drop of the temperature to normal. A similar experience was noted by Dr. H. E. Pearse.

Imbecility seems to be due either to *maldevelopment* or to *atrophy*, the latter being the result of any one of a considerable number of affections, such as meningo-encephalitis, embolism, hæmorrhage, pressure from old cysts, etc. In atrophic cases we have *destruction* of cerebral tissue by a process of fatty degeneration, or by the formation of sclerotic or scar tissue, which destroys and supplants the normal structures in a manner similar to that in which we know cirrhosis destroys the parenchyma of the kidney in chronic interstitial nephritis. If during the active stages of this atrophic process operation could arrest its progress, then much good might be done, but, unfortunately, in many of these cases such a conclusion can hardly be hoped for. Where the atrophic changes are not far advanced and are excited and kept up by the irritation of a cyst, or such like lesion, or even perhaps by the presence of meningitis, then early operation may prevent further atrophy. It is, however, inconceivable that such treatment or any treatment could restore the already destroyed nerve-cells to functional activity. When the atrophic condition has once been fully developed, then operation presents no prospects of good resulting, unless we think that, in individual cases, sufficient potential tissue remains to be of service, if put under conditions in which development is possible. In this connection it is worth noting that the brain can develop at least up till the

twentieth year.¹ For these reasons it is probable that, even had Case I survived the operation, no great improvement would have been obtained.

So much for the atrophic causes of imbecility, but idiocy may be the result of maldevelopment of several classes, only two of which will be considered here. Of microcephalia nothing may be said. It is outside the scope of this paper.

I. *Simple Maldevelopment*.—To this class belongs Case II, and also a case I saw in consultation with Drs. Griffith and Lanphear, and in which these surgeons operated with very gratifying results.² Here we have a condition of the brain in which there is “an *apparent* (the italics are mine) atrophic condition of the minute structures of the hemisphere, chiefly cortical, the cells resembling those of a new-born child, but with no apparent gross defects in the brain.”³ In cases belonging to this class, we may hope to get the best results; in fact, already, with our imperfect knowledge and probably imperfect technique, the results have been most astonishing. To me it was a most wonderful sensation to hear a boy, only a few hours after he had been operated on, ask distinctly and *intelligently* for water when he had never been known to ask for even such a simple thing as that throughout the whole of his previous existence.

Craniotomy in this class of cases seems to remove a *something* which has been hindering development, and to a greater or less extent impeding or preventing the action of some parts of the brain which have already been more or less developed. The fact that in such cases when the skull is first opened there has always, so far as my observation goes, been a total lack of the normal cerebral pulsation, and that this pulsation appeared after a short time, points to a condition of exaggerated tension as the cause of the maldevelopment. Our knowledge is, however, too limited as yet to dogmatize. As already mentioned, improvement of mental function is observable within a few hours of operation. This change is, of course, too early to be explained

¹ M. Allen Starr, *Brain Surg.*, p. 134.

² *Proc. Mo. State Med. Asso.*, 1893.

³ Starr, *loc. cit.*

by any new cerebral development; it must be due to the removal of some impediment which has been keeping already existing cerebral substance in a state of functional inactivity.

II. *Porencephalia* is another form of maldevelopment occasioning imbecility. Here the agenesis is so great that an opening exists between the surface of the brain and a widely-dilated lateral ventricle beneath, the ventricle and the opening into it being full of fluid. This condition is very similar to hydrocephalus, but there is no distention of the head itself.

I have only had the opportunity of studying one case of porencephalia. In this case the symptoms were entirely similar to those seen in simple maldevelopment. Dr. Griffith, with whom I saw the case, operated. During the course of the operation the dura was incised, a great quantity of cerebro-spinal fluid escaped, and the whole hemisphere seemed to collapse. The patient died the same day. At the autopsy the brain was found to be unilaterally porencephalic. The same surgeon has related to me an exactly similar case in which the result of operation was the same. Dr. Clayton Parkhill, of Denver, recently told me that he had performed craniotomy on one side of an idiot's skull, abstaining from making any dural incision. No benefit accrued from the operation. When the other side of the skull was opened fluid was seen under the dura, and on an incision being made much clear fluid escaped, and the mere shell of tissue which represented the hemisphere collapsed. Death soon ensued.

Such experiences show that it is certainly at the present day impossible to make a differential diagnosis between simple maldevelopment and porencephalia, but that when the skull has been opened, and examination of the meninges indicates the latter lesion, then the membranes should not be incised. If it is thought that the withdrawal of a small quantity of the fluid will be of benefit this can be done with a hypodermic needle, but any rapid escape of the fluid is extremely hazardous.

In selecting cases for operation the patient's personal history must be our main guide. Careful inquiries must be made as to the presence or absence in the past of symptoms which would

lead us to suspect that hæmorrhage, inflammation, or such like troubles had been the original cause of the imbecility. A point in the history which I consider of very great importance, but of which, so far as I know, no notice has been taken, is the question whether or not before the imbecility manifested itself there was any mental development, and whether the imbecility was accompanied by the loss of the mental development previously attained, or merely by this development ceasing to progress.

In non-microcephalic cases where up to a certain time mental development had progressed but subsequently has been lost, we are surely justified in believing that an atrophic process has been established, and if it has lasted any considerable time, but little may be expected from operation. On the other hand, if, when imbecility has been established, this degree of development has not been lost, but has remained *in statu quo*, or has advanced even infinitesimally, we are forced to the conclusion that the trouble is not atrophy but maldevelopment, and operation *may* here provide the conditions necessary for proper progress. If there has never been any mental development exhibited, we have no means, other than exploration, of knowing the cause of the idiocy.

In microcephalia the conditions are different, but, to quote Kipling, "that is another story."

When performing the operation of craniotomy the surgeon ought to remove a wide strip of bone. In a case of congenital microcephalia, in which it may be incidentally stated that no improvement resulted, I had occasion to operate twice on the same side of the head. Although only a few weeks divided the two operations, yet at the second setting it was found that very much of the opening formerly made had been filled up by a new formation of bone. This was so marked that it gave one the impression that a defect one-half inch in width would soon have been obliterated.

